Indiana's Universal Newborn Hearing Screening Program

Why Universal Newborn Hearing Screening?

- UNHS has become the standard of care with 34 states participating in this program. The American Academy of Pediatrics, as well as other hearing health organizations, advocate for universal newborn hearing screening.

- Technology now exists to provide safe, cost effective and reliable methods to assess hearing in newborns, making UNHS possible and practical. Screening involves the use of non-invasive, objective physiologic measures.

- Screening procedures for newborns can lead to detection of significant bilateral or unilateral hearing loss. **All children can be evaluated for hearing loss, regardless of their age.**

- Hearing loss is invisible, but the effects can lead to lack of exposure to language and can cause lifelong cognitive, educational and vocational challenges. **This is preventable with early intervention and family education.**

- **With 1 out of every 300 infants born with significant hearing loss, it is one of the most common health conditions found in newborns.** In Indiana approximately 250-300 babies will be identified annually.

- If only high-risk babies were screened, half of all babies with hearing loss would be missed.

- The incidence of congenital hearing loss is greater than the sum total of all other conditions detected by newborn metabolic blood screening tests.

- More than 90% of infants who are born with or develop early onset hearing loss have parents and families with normal hearing.

- **Without universal newborn hearing screening, the average age of diagnosis is over two years of age.**

- Recent research indicates that children identified with hearing loss, who receive intervention before six months of age, develop language (spoken or signed) comparable with their hearing peers.

- **Don't wait for signs of hearing loss to appear.** Many children with hearing loss will not have an obvious speech/language delay until 2,3, or even 4 years of age.

The information above was compiled from numerous sources on UNHS including ASHA, Pediatric News, American Academy of Pediatrics, SHHH, and the National Campaign for Hearing Health. 2-19-2002

Universal Newborn Hearing Screening Best Practice

1st Screen

Pass – both ears
Parent receives certificate – passed

Did not pass – one or both ears
2nd Screen is done

Pass-with risk indicators
Parent receives certificate and referral for testing at 6-9 months

Pass both ears

Refer in one or both ears

Did not pass – one or both ears
Parent receives certificate – referral
What to tell parent – *"Your child did not pass hearing screening. This does not mean he has a hearing loss. We do a hearing screening. We are referring your baby for a diagnostic test. The diagnostic test will clarify how your baby is hearing. It is important that you follow up as soon as possible with the hearing diagnostic test. Early detection of hearing loss is important for language development."*

Refer per hospital policy to: Physician, Audiologist, and/or First Steps
First Steps

Complete blood spot card for all babies
Report to ISDH on the Monthly Summary Report (MSR):

1. Any baby not receiving UNHS
2. Babies that don't pass UNHS
3. Babies that pass but are at risk for delayed onset of hearing loss
    a. Family history of permanent childhood hearing loss.
    b. Congenital infection (CMV, rubella, herpes, syphilis, toxoplasmosis).
    c. Hyperbilirubenemia requiring exchange transfusion.

ISDH Fax #: (317) 234-2995

Tips for UNHS Screeners
Screening Procedures

OAE & AABR: (Natus/AUDX/Echoscreen)

Testing should be done when the baby is quiet (preferably sleeping) and relaxed, well fed and comfortable.

Swaddling the infant often helps.

Testing area should be quiet (avoid talking, ringing phones, running water)

Confirm the ear to be tested.

OAE: (AUDX/Echoscreen)

Visually inspect the ear canal for debris (wax, blood, vernix).

Seat the earphone probe by gently pulling the ear up and out: this will open up the canal.

Begin the test once the probe is placed and baby has quieted.

If the baby does not pass on the first try,

- Remove the probe and check for debris
- Replace the tip if needed
- Clean probe if needed
- Reposition the probe and repeat the screen

Best practice: Wait a few hours before repeating the screen.

Adapted from UNHS sources from ID.

Tips for UNHS Screeners
What to tell Parents

1. **Prior to screening:** Give them the ISDH brochure explaining the UNHS program and tell them this is one of the newborn screens required for all babies.
2. **If the baby passes:** Tell them that their baby passed the hearing screening. Give them the ISDH certificate. Show them the checklist for language and hearing development milestones on the certificate and remind them to monitor the child's progress and development.
3. **If the baby passes but has risk factors:** Tell them that their baby passed the hearing screening but may be at risk for delayed onset hearing loss. Give them the ISDH certificate. Show them the section about what causes infant hearing loss, as well as the language and hearing development milestones. Discuss a referral to the PMP and First Steps for follow-up testing at 9-12 months of age.
4. **If the baby does not pass:** Give them the ISDH brochure for referral. Do not use the words "failed" or "deaf". Tell them that their baby will need further testing. Keep what you say simple. Reassure the family that there are many reasons why this might happen and that diagnostic testing will clarify the baby's hearing status and should be completed in a timely manner. Inform them about a referral to their PMP and First Steps for help in both scheduling appointments and financial assistance. Be supportive about follow-up in a timely manner.

Adapted from UNHS sources from ID.

Tips For UNHS Screeners
Minimizing Referral Rates

Test while the baby is quiet, relaxed (preferably sleeping), well fed and comfortable. Swaddling the infant often helps.

If a second screen is necessary, wait a few hours. This can significantly reduce the referral rates. Always rescreen both ears.

Screening will be faster and more effective if you minimize noise and distraction before screening. Testing area should be quiet (avoid talking, ringing phones, running water, etc.).

It is important to have a backup equipment plan in the event of a breakdown. *See Tips for Screeners / Back-up Equipment for suggestions on a plan.

OAE specific tips:
For OAE screening, the single most important factor in reducing referral rates is achieving a good probe fit:

- Visually inspect the ear canal for debris (wax, blood, vernix)
- Seat the earphone probe by gently pulling the ear up and out: this will open up the canal
- Begin the test once the probe is placed and baby has quieted
- If the baby does not pass on the first try,
    - Remove the probe and check for debris
    - Replace the tip if needed
    - Clean probe if needed
    - Reposition the probe and repeat the screen

Adapted from UNHS sources.

Tips for Blood Spot Card Completion

Instructions for UNHS Portion of Blood Spot Card

1. **Every effort should be made to have the UNHS results as well as the blood spot sample entered on the blood spot card before it is sent to the lab.** A complete set of data will be entered into the system for each infant.
2. To facilitate data entry, the UNHS screening should be completed and entered on to the blood spot card immediately upon completion. Complete the information requested
    a. Check the appropriate box for initial (one screen and passes both ears) or re-screen (if the baby requires a second test for one or both ears)
    b. Enter the date of reported screen
    c. Indicate whether pass or refer for each ear
    d. If risk factors are present please indicate (First Step risk factors include: family history/congenital infection/hyperbilirubinemia requiring exchange transfusion)
    e. If not screened, check the appropriate box (Deceased/Transferred/Hospital error/NICU/Unauthorized Refusal/Religious Refusal/Equipment Problems/ Other)
3. **Do not delay in sending the blood spot card** if hearing screening is not completed. All newborn screen blood samples should be sent within 24 hours of collection, even if the UNHS screen has not been done. A delay in sending the blood spot info could result in a delay in diagnosis.
4. If hearing screening has not been completed (due to transfer/NICU/etc.) retain the pink "hearing" pull out sheet and keep until hearing screen is completed. When the hearing screening has been completed, enter the information on the copy and forward to the IU lab for data entry. Newborn Screening Laboratory, P. O. Box 770, Indianapolis, IN 46206
5. Continue to complete the MSR packet each month and send it to ISDH as usual.

UNHS Missed Baby Alert!

When completing the Monthly Summary Report:

1. Note all missed babies on the MSR.
2. Have a standard letter ready and mail to the baby's family and the baby's physician stating the importance of the screen and the need for the family to return to the hospital for this screening prior to 14 days of age.
3. If the family does not return for follow-up contact the Nurse Consultant at ISDH for assistance. This will allow for quicker intervention for the babies who did not receive a screen prior to discharge.

Tips
Every Baby Screened before Discharge

1. Work with the hospital's information systems department. Develop a protocol to have a list of every baby born at your facility. Cross check this list daily with your UNHS log.
2. Consider making UNHS a standard of care for all infants born in your hospital.
3. Routinely screen all infants using a standard timetable (e.g. 6-12 hours of age, or at designated times during the day to establish a routine for the staff).
4. Have a system in place at your facility to be notified when a baby is discharged from the NICU to the well-baby nursery so that hearing screening can be completed.
5. Include hearing screening in pre-natal classes so parents understand the importance of the screen being completed prior to discharge.
6. If a baby is inadvertently discharged without a hearing screening, notify the family and the primary care physician, so that the family can return for a screen prior to the baby being 14 days of age.
7. Maintain good records and report complete data to ISDH.

TIPS FOR SCREENERS

Back-up Equipment

If your hearing screening equipment malfunctions, a back-up plan needs to be in place in order to avoid missing babies or having to bring babies back after discharge for testing.

Possible suggestions include:

1. Many hospitals have "sister" facilities that may be able to loan equipment to each other.

2. Some manufacturers offer loaner equipment—arrangements can be made for equipment to be sent immediately for loan until repairs are completed. Check with your manufacturer's sales representative regarding this possibility.

3. Check with local audiologists or ENT practices to see if they have equipment that they could loan or contract to use for a small fee.

If the equipment malfunctions, have a plan in place for a back-up unit. Babies who miss the screening due to equipment problems must be brought back for screening when the equipment is repaired. Being prepared for equipment problems will decrease the delays in screening all of your newborns.

Tips for the Person Who Completes the MSR for UNHS

1. Make sure all babies are screened prior to discharge.
2. Make sure all babies who need a second screen are screened prior to discharge.
3. Avoid "extra" screens to have a baby pass.
4. Have a back-up plan in place should you have equipment malfunction.
5. If a baby is "missed" (not screened prior to discharge) contact the family to return for an outpatient re-screen promptly. Indicate these babies on the MSR report (1I.) and Reason Code Sheet (1I.)
    a. If the baby returns for screening, notify the Chief Nurse Consultant, Bess Godard, of the results. Contact her at 317 233-1266 or by email at bgodard@isdh.state.in.us. Also include those babies on the next months Reason Code Sheet under walk-ins (1D.)
    b. If the baby does not return for screening, notify the Chief Nurse Consultant, Bess Godard so that ISDH can follow-up with a letter to the family in a timely manner. Contact her at 317 233-1266 or by email at bgodard@isdh.state.in.us.
    c. Remember, an older baby is more difficult to screen.
6. Transferred babies are a "shared" responsibility. Please keep track of infants who are transferred out prior to screening and include them on the Reason Code Sheet (d.). Follow-up with the receiving hospital to make sure the transferred infant was screened and document this in your records. A new form to help track transferred babies is now available. The receiving hospital should return this form to ISDH and it can also be used to report back to the birthing hospital for their records. The birthing hospital will need to follow-up any babies who were transferred for whom they receive no information.
7. Remember to complete the Infants Referred for Follow-up for all babies you refer and indicate by the code what type of referral was made. This includes infants referred for not passing two screens and those with the three risk factors.
8. Please list the names of babies in the NICU on the reason code page, if they are not screened within the first month. It will assist ISDH in locating these babies in the future should they be discharged without a screen.
9. Check your numbers prior to sending in the MSR. If you have 10 babies on line 1I, there should be 10 babies on the Reason Code Sheet. Make sure you code why those babies are on the list. If you have 2 infants on line 2A (At risk) there should be 2 infants listed on the Follow-up Sheet under Codes D, E, or F. If you have 3 babies listed on 3A. (Not Passing), there should be 3 babies listed on the Follow-up Sheet under Codes A, B, or C.
10. If parents sign a religious waiver, make sure you attach a copy to the MSR report.
11. Refer to the instructions provided with the report form for any questions you have or contact the Chief Nurse Consultant at 317 233-1266 or by email at bgodard@isdh.state.in.us.

Tips for UNHS Screeners
Referral for Delayed Onset Risk Factors

Indiana's UNHS Policy Manual identifies three risk factors for delayed onset hearing loss that require referral to Indiana State Department of Health and First Steps Early Intervention programs. Babies who pass the screening but have one of the following risk factors need to be referred. Risk factors include:

1. A family history of permanent childhood hearing loss
2. Exposure to infection before birth, such as herpes, syphilis, cytomegalovirus (CMV) and toxoplasmosis
3. Bad jaundice (hyperbilirubinemia) that needed a special procedure (exchange transfusion)

Families of babies who pass the screening, but are identified as having one of the above risk factors, should:

1. Be informed about the risk factors identified
2. Be made aware of the hearing and language developmental milestones on the screening certificate that is given to the family and told to monitor the child's progress
3. Be referred to their PMP on the discharge summary
4. Be referred to ISDH on the Monthly Summary Report and to First Steps, using the form in the UNHS Policy Manual for follow-up evaluation at 9-12 months of age and every six months after until age 3
5. Sign the reciprocal release of information enabling the Newborn Screening Department at the Indiana State Department of Health to receive copies of follow-up testing
6. Be informed of the results and the importance of follow-up testing

Tips for UNHS Screeners
Referral for Other Risk Indicators

Other risk factors do exist (as identified below) and may be present. Babies who pass the screen but have one of these additional risk factors, should be referred to their primary care physician. A referral to the child's primary care physician should be completed on the discharge summary.

1. Spent more than 48 hours in the Neonatal Intensive Care Unit (NICU)
2. A condition known to be associated with hearing loss (genetic/syndrome)
3. Head, face or ears that are formed in a different way than usual (craniofacial anomalies)
4. An infection around the brain and spinal cord caused by bacteria (bacterial meningitis)
5. Parent or care giver concern regarding the development of hearing and language

Families of babies who pass the screening, but are identified as having one of the above risk factors, should:

1. Be informed about the risk factors identified
2. Be referred to their PMP on the discharge summary for follow-up testing
3. Be made aware of the hearing and language developmental milestones on the screening certificate that is given to the family and told to monitor the child's progress
4. Be informed of the results and the importance of follow-up testing

Newborn At-Risk Criteria for Delayed Onset/Progressive Hearing Loss

State-Mandated for follow-up

- **Family History of Congenital Childhood Hearing Loss**
  - Does not include history of middle ear infections/tubes
  - Does not include family members with know causes of hearing loss like Rubella, Meningitis, or loud noise exposure & trauma
  - Does include family members with hearing loss in one or both ears since childhood from unknown cause or history of genetic hearing loss

- **Maternal Infection (TORCH)** includes:
  - **Toxoplasmosis** – infected during or just before pregnancy, especially 1st trimester
  - **Group Beta Strep (GBS)** – untreated and baby is delivered vaginally
  - **Syphilis** – infected during pregnancy, baby can be treated prior to delivery
  - **Rubella** – infected primarily during the first trimester
  - **Cytomegalovirus (CMV)** – can be transmitted through the placenta, birth canal or postnatally through breast milk
  - **Herpes Simplex Virus (HSV)** – active infection during pregnancy , primarily active during vaginal delivery

- **Hyperbilirubinemia (Jaundice)**
  - At levels exceeding indication for exchange transfusion

Other at-risk factors for hearing loss in infants exist and would routinely be investigated by the infant's primary care physician. These factors include:

- Craniofacial abnormalities
- Syndromes that are commonly associated with hearing loss, such as, Down's Syndrome, Usher Syndrome, Waardenburg Syndrome, and Neurofibromatosis Type 2, to name a few
- Low birth weight below 3.3 lbs.
- Prolonged ventilation (> 10 days)
- Aminoglycosides (>5 days) and loop diuretics used in combination with aminoglycosides
- Apgar scores of 0-3 at 5 minutes or those who fail to initiate spontaneous respiration by 10 minutes or those w/ hypotonia persisting to 2 hrs. of age
- Any infant not passing two newborn hearing screenings
- Parental concern